

transposition. Yet all the patients that they studied with atrioventricular discordance also had ventriculo-arterial discordance. As far as we are aware (and we are supported by Metcalfe and Somerville¹) this combination is the one usually referred to as congenitally corrected transposition. It is quite clear that our comment was made specifically in the context of atrioventricular discordance. Thirdly, they accuse us of raising the equiplanar insertion of the atrioventricular valve leaflet to the status of a pathognomonic sign of an inlet ventricular septal defect. We do not know how they reached this conclusion. This statement is not to be found anywhere in our writings. Indeed, in the introduction to the paper they refer to, we stated—alluding to the valve attachments being at the same level—“This finding is of course characteristic of a double inlet atrioventricular connexion, but in this case its importance is trivial compared to the other abnormalities present.” There is another equally important reason why we would not make this statement. The offsetting of the atrioventricular valve leaflets and the presence of a muscle bar projecting downwards from the atrioventricular junction to roof a defect clearly differentiates an inlet muscular defect from a perimembranous inlet defect.

We are grateful to take this opportunity to clarify our figure legends in the four instances in which pathological specimens were shown with the corresponding echocardiograms. As these stand in the text they could lead to some confusion. In Fig. 3 and Fig. 4 the pathological specimens were not from the patient whose echocardiogram is shown but were

sections from similar patients drawn from the cardiopathological collections of the three centres concerned. In Fig. 9 and Fig. 10 the pathological specimens were from the patient whose echocardiogram is shown.

Finally we wish to defend the principle of using corresponding anatomical specimens to illustrate echocardiographic stop frame images. This has now become a widely adopted form of presentation, and such correlative papers have appeared in the majority of leading cardiology journals. Indeed a large number of such papers have undergone review and publication in the *British Heart Journal*. We feel strongly that such a correlative presentation leads to a greater appreciation of the information which can be derived from echocardiographic images obtained from patients with complex heart disease. We would further suggest that Foale *et al* are in the minority in voicing criticisms of what is now a widely accepted form of correlative presentation.

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Reference

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Myocardial disorganisation in hypertrophic cardiomyopathy

Sir,

“Whatever is only almost true is quite false, and among the most dangerous of errors, because being so near truth it is the most likely to lead astray.”

I was reminded of the words of Henry Ward Beecher when reading Dr Maron's editorial in the July issue of this journal (1983;50:1–3). In his rebuttal of our work¹ Dr Maron makes errors not only in misunderstanding what we had to say but also in pointing in the wrong direction when complaining that others have generated a controversy regarding the role of myocardial disorganisation (or disarray). Indeed, Dr

Maron does not hesitate to extrapolate from his interpretation of the works of others, including the paper by Becker and Caruso, that those investigators lack any meaningful experience with hypertrophic cardiomyopathy.

Regarding the issue at stake it may be necessary to briefly reiterate the most salient points. Firstly, the rather loose use of terms such as “characteristic,” “typical,” and “specific,” when referring to myocardial disorganisation in cases with hypertrophic cardiomyopathy, has led to the misconception that this particular histological feature is a useful diagnostic

marker in a clinical setting. The problem started after the excellent article by Ferrans and associates,² concerned in particular with the pathogenesis of the disease, stating that disarray was "unique" for hypertrophic cardiomyopathy. The statement implied that disorganisation was a pathognomonic feature and hence could serve as a diagnostic marker. Subsequently, the Bethesda group—with Dr Maron quite often in the lead—gradually shifted their stand using phrases such as "not absolutely specific"^{3,4} and "not specific and commonly found in other forms of myocardial hypertrophy."⁵ They presently claim that myocardial disarray is a highly sensitive and specific marker for hypertrophic cardiomyopathy when sites of naturally occurring "disorganisation" are excluded. Their diagnostic sections are taken midway through the ventricular septum or from the affected left ventricular free wall, in through and through fashion, and then microscopic sections are sliced in a transverse plane.⁶⁻⁸

In the light of these statements we have simply questioned the relevance of myocardial disarray as a diagnostic marker in a clinical setting.¹ This possibility has not passed unnoticed to these distinguished investigators, since they also had reached the conclusion that small pieces of myocardial tissue are of limited value in diagnosing hypertrophic cardiomyopathy.^{9,10}

In the light of the above it is sad that Dr Maron in his paragraph "cardiac muscle cell disorganisation as a marker for hypertrophic cardiomyopathy" fails to point out that the quantitative assessments to which he refers are based on sections encompassing the full thickness of the ventricular septum or the left ventricular free wall. His statement, therefore, that myocardial disarray distinguished hypertrophic cardiomyopathy from other lesions producing left ventricular hypertrophy should be considered with this restriction in mind. It is here that Dr Maron comes quite close to the truth but for reasons unknown has decided not to go all the way. This is the more regrettable since we agree that full thickness sections will usually show disarray as the leading histological feature. As far as I am aware all other workers in the field will agree with this statement. The point we made, and which Dr Maron apparently has not appreciated, is that small biopsy specimens taken from the left ventricular endomyocardium are unlikely to be diagnostic for hypertrophic cardiomyopathy since in that circumstance myocardial disarray is a poor marker. On that basis we question whether myocardial disorganisation has a place in the clinical decision making when a patient is suspected of having this disease.

Dr Maron has apparently not appreciated the fact that we studied normal hearts purposely in order to see whether or not disorganisation occurred in the

setting of the normal heart and to what extent it could affect the interpretation of tissue samples of normal myocardium. Since our study showed that "disorganisation" could be produced by changing the orientation of the plane of section we cautioned against an overenthusiastic diagnosis of myocardial disorganisation, with all its implications, based on routinely processed histological sections. We had the impression that this was all clear and straightforward, not giving rise to further bias. To our surprise *Cardiology* 1983 contains a similar misinterpretation of our work.¹¹ Be that as it may, in our paper¹ nowhere did we extrapolate our findings to hypertrophic cardiomyopathy so as to suggest that myocardial disarray is *not* a feature of that disease. I challenge Dr Maron to quote from our paper,¹ without distorting the context, any sentence that even suggests this implication. It seems, sir, that this interpretation is based on pure inference on the part of Dr Maron rather than on any implication by ourselves.

As far as I am aware none of the workers quoted by Dr Maron, including Becker and Caruso, has claimed that myocardial disorganisation is *not* a feature of hypertrophic cardiomyopathy. Indeed, when Dr Maron reads and digests our paper fully he will undoubtedly encounter a crisp sentence: "The above discussion should not be construed as suggesting that myocardial disarray is not a feature of hypertrophic cardiomyopathy" (p 536). It is ironic, therefore, that I agree wholeheartedly with Dr Maron's final conclusion that further study of myocardial disarray may be beneficial in understanding the natural history of hypertrophic cardiomyopathy.

I am left with the overall conclusion that Dr Maron in his editorial has in no way aired another viewpoint. Instead, he has produced still further obfuscation with his "straw man."

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Notices

"Congenital heart disease made simple"

A course on the major congenital cardiac anomalies is to be held from 8 to 11 October 1984 at the Institute of Child Health, London. Further information may be obtained from: Dr S G Haworth, Institute of Child Health, 30 Guilford Street, London WC1N 1EH.

British Cardiac Society

The Autumn Meeting in 1984 will be held on 3 and 4 December 1984, and the closing date for receipt of abstracts will be 15 August 1984.